

## PAPER

# Removal of large acoustic neurinomas (vestibular schwannomas) by the retrosigmoid approach with no mortality and minimal morbidity

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*J Neurol Neurosurg Psychiatry* 2004;**75**:453–458. doi: 10.1136/jnnp.2003.010827

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Received 21 January 2003  
In revised form  
23 April 2003  
Accepted 28 June 2003

**Objective:** To evaluate the safety and efficacy of removing large acoustic neurinomas ( $\geq 3$  cm) by the retrosigmoid approach.

**Methods:** Large acoustic neurinomas (mean (SD), 4.1 (0.6) cm) were removed from 50 consecutive patients by the retrosigmoid suboccipital approach while monitoring the facial nerve using a facial stimulator-monitor. Excision began with the large extrameatal portion of the tumour, followed by removal of the intrameatal tumour, and then removal of the residual tumour in the extrameatal region just outside the porus acusticus. The last pieces of tumour were removed by sharp dissection from the facial nerve bidirectionally, and resected cautiously in a piecemeal fashion.

**Results:** There were no postoperative deaths. The tumour was removed completely in 43 of 50 patients (86%). The facial nerve was anatomically preserved in 92% of the patients and 84% had excellent facial nerve function (House-Brackmann grade 1/2). One patient recovered useful hearing after tumour removal. Cerebrospinal fluid leak occurred in 4%, but there were no cases of meningitis. All but two patients (96%) had a good functional outcome.

**Conclusions:** The method resulted in a high rate of functional facial nerve preservation, a low incidence of complications, and good functional outcomes, with no mortality and minimal morbidity. Very favourable results can be obtained using the retrosigmoid approach for the removal of large acoustic neurinomas.

Surgical removal is the sole treatment for large acoustic neurinomas because stereotactic radiosurgery is not applicable.<sup>1,2</sup> The translabyrinthine,<sup>3,4</sup> retrosigmoid suboccipital,<sup>5-7</sup> and middle fossa approaches<sup>8,9</sup> are the three basic approaches for the removal of these tumours. Recent papers have recommended the translabyrinthine approach for the removal of large tumours and have reported good results with this technique.<sup>4,10-12</sup> However, we have used the retrosigmoid suboccipital approach for over 20 years for the removal of acoustic neurinomas of all sizes. In this study, we reviewed 50 consecutive patients with large tumours that were removed by the retrosigmoid approach over the past 10 years.

## METHODS

### Patient population

The subjects consisted of 89 consecutive patients with unilateral acoustic neurinomas in the period from 1992 to 2001. The extrameatal diameter of the tumours was  $\geq 3$  cm in 50 of the 89 tumours (*large*; 56%), and  $< 3$  cm in 39 (*small to medium*; 44%). In this study we analysed the 50 patients with large tumours. Patients with neurofibromatosis type 2 or with a recurrent tumour were excluded from the study.

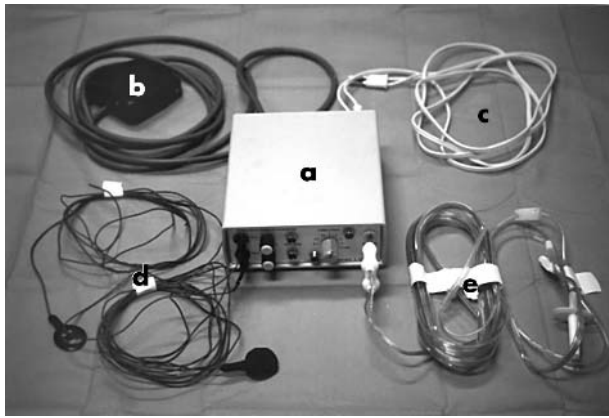
### Surgical procedure

All operations were done by single neurosurgeon (IY), using the retrosigmoid suboccipital transmeatal approach in the lateral position. A curvilinear skin incision of 12 cm length was placed 3–4 cm posterior to the mastoid. A longer incision placed more medially was used in patients with short thick necks, while a shorter skin incision placed more laterally was used in those with long thin necks. Irrespective of the tumour size, a retrosigmoid suboccipital craniotomy of 5 cm length and 4 cm width was made to expose the posterior part of the

sigmoid sinus and the inferior part of the transverse sinus. The foramen magnum was not usually opened.

The microsurgical removal of the tumours comprised three steps: cytorreduction of the large extrameatal cerebellopontine angle portion of the tumour, the removal of the intrameatal tumour, and the removal of the last lump of tumour remaining in the extrameatal region just outside the porus acusticus. After confirming that the facial nerve was not on the dorsal surface of the cerebellopontine angle tumour, using a facial stimulator-monitor designed by us,<sup>13</sup> internal decompression of the large extrameatal tumour was undertaken with an ultrasonic surgical aspirator (CUSA EXcel, Vallylab, Boulder, Colorado, USA). We used the aspirator cautiously so as not to rupture the tumour capsule or injure normal structures on the capsule. Dissection of the reduced cerebellopontine angle tumour from the brain stem in a caudal to rostral direction identified the root entry/exit zone of the facial and acoustic nerves. Resection of the posterior wall of the internal auditory meatus was minimised to prevent related complications, including cerebrospinal fluid (CSF) leak and facial nerve injury.<sup>14</sup> The last lump of tumour in the extrameatal region just outside the porus acusticus adhered to the facial nerve most tightly.<sup>6</sup> The tumour and facial nerve were dissected bidirectionally. The last piece was resected by sharp dissection in a piecemeal fashion using microscissors and a knife, while confirming the maintenance of facial nerve function with the stimulator-monitor. No bipolar coagulation was applied for haemostasis in this region.

The internal auditory meatus was reconstructed to prevent CSF leaks and to regain the CSF space in the internal auditory meatus. This surgical technique has been published elsewhere.<sup>15</sup> After watertight dural closure, we did a cranioplasty using an autobone or artificial bone (Biobone, Johnson & Johnson, Tokyo, Japan).



**Figure 1** Facial nerve stimulator-monitor arranged in a circuit with a bipolar coagulator: (a) main box, (b) foot switch, (c) cord connecting the stimulator with a bipolar coagulator, (d) a two channel detector for the facial muscle movement, (e) a pair of bipolar forceps connected to the stimulator using the cord of a bipolar coagulator.

Intraoperative facial nerve monitoring was done routinely using the facial nerve stimulator-monitor.<sup>13</sup> Facial muscle movement in response to the electrical stimulation was detected by a two channel contraction detector built into the apparatus (fig 1).

#### Follow up in the outpatient clinic, outcome measures, and statistical analysis

After tumour removal, all patients were followed up in our outpatient clinic regularly every six months and underwent magnetic resonance imaging (MRI) once a year. One year after the tumour removal, facial nerve function and functional outcome were assessed. Facial nerve function was evaluated using House-Brackmann (H-B) grades,<sup>16</sup> and was categorised as *excellent* (H-B grade 1/2), *intermediate* (H-B grade 3/4), or *poor* (H-B grade 5/6). Functional outcome was measured using the Karnofsky scale. Data were analysed using a statistical software application (Statview, SAS Institute Inc, Cary, North Carolina, USA). Descriptive data are expressed as mean (SD). A probability (*p*) value of <0.05 was considered significant.

## RESULTS

### Preoperative clinical signs

Table 1 summarises the clinical characteristics of 89 unilateral acoustic neurinomas. There was no significant difference in the age, sex, and side of tumour between the patients with large (*n* = 50) or small to medium tumours (*n* = 39). Three of 15 patients with hydrocephalus presented with symptoms of normal pressure hydrocephalus—that is, gait disturbance, dementia, and incontinence. Although unilateral hearing loss was the most common sign in the patients with large acoustic neurinomas, two patients had normal hearing. Twenty two per cent of patients had useful hearing, which we defined as *class A* or *class B* according to the guidelines of the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology, Head and Neck Surgery Foundation (table 2).<sup>17</sup>

### Preoperative magnetic resonance imaging

All patients underwent preoperative MRI (T1 weighted imaging (T1–WI) before and after gadolinium (Gd) administration, and T2–WI). The extrameatal diameter of the large tumours ranged from 3.0 to 5.8 cm (mean (SD), 4.1 (0.6) cm); 25 tumours (50%) were 3.0–3.9 cm in size, 20 (40%) were 4.0–4.9 cm, and five (10%) were ≥5.0 cm. The tumour

consistency was classified as *solid*, *partly cystic*, or *mostly cystic* using T1–WI after Gd administration (table 3). Cyst formation was significantly more common in the large tumours than in the small to medium tumours. Two patients presented with severe headache of sudden onset, and showed a massive haemorrhage in the tumour cyst on MRI. Five patients presented with severe ataxia, and showed peritumoral oedema extending to the middle cerebellar peduncle on T2–WI.

### Completeness of tumour removal

The completeness of tumour removal was judged by surgical records and postoperative MRI. Tumour removal was complete in 43 patients (86%) and subtotal in seven. All seven subtotal removals were done in the early study period (1992 to 1996). The proportions of cases with subtotal/total removal were 7/27 (26%) and 0/23 (0%) in the periods from 1992 to 1996 and from 1997 to 2001, respectively. The difference between the two periods was significant (*p* = 0.01, Fisher's probability test). Removal was subtotal in the following: in three patients who requested subtotal removal rather than risk facial palsy, so a small part of the tumour adhering to the facial nerve was intentionally left; in two patients in whom a small part of the tumour adhering to the brain stem was left because hypertension and tachycardia occurred when we attempted to dissect the tumour; and in one patient in whom intraoperative cerebellar swelling resulted in subtotal removal.

### Facial nerve outcome

The facial nerve was anatomically preserved during the tumour removal in 46 patients (92%). Of four patients in whom the facial nerve was divided, three underwent primary nerve anastomosis using a nerve graft and one underwent hypoglossal–facial nerve anastomosis. These facial nerve reconstructions resulted in intermediate facial nerve function. Of 46 patients with anatomically preserved facial nerves, 42 (84%) showed excellent facial nerve function, three showed intermediate function, and one showed poor function (fig 2).

### Postoperative complications and functional outcome

There were no postoperative deaths. Cerebellar haemorrhage resulted in persistent ataxia in two patients. A supratentorial epidural haematoma occurred in one patient who underwent ventricular drainage for hydrocephalus. Two patients developed CSF leaks, which were treated by lumbar drainage only, and no surgical repair was necessary. There were no postoperative cases of meningitis. Preoperative trigeminal neuropathy improved or resolved completely in most patients after tumour removal. Hypaesthesia of the face was exacerbated in three patients. Useful hearing was not preserved in 49 patients; however, in one patient tumour removal improved the hearing from class C to class B, and useful hearing was restored (fig 3). Endotracheal intubation during surgery caused bilateral recurrent nerve palsy in two patients, resulting in transient hoarseness and swallowing disturbance.<sup>18 19</sup> One patient developed dyspnoea from neck swelling and needed a temporary tracheostomy.

Functional outcome was good in 48 patients (Karnofsky score, 90). Two patients had Karnofsky scores of 80 and 70 because of persistent ataxia. In five patients tumour removal caused resolution of severe preoperative ataxia as well as of the peritumoral oedema (fig 4). Of 15 patients with preoperative hydrocephalus, five (33%) required CSF diversion.

**Table 1** Clinical characteristics of 89 unilateral acoustic neurinomas

	Large tumour (≥3 cm)	Small-medium tumour (<3 cm)	p Value*
No of patients	50	39	
Age (years) (mean (SD), range)	52 (15), 8 to 73	51 (11), 24 to 69	NS
Sex (male:female)	21:29	21:18	NS
Side of tumour (right:left)	22:28	19:20	NS
<i>Clinical signs and symptoms</i>			
Hydrocephalus	15 (30%)	1 (3%)	p<0.01
Papilloedema	5 (10%)	1 (3%)	NS
Trigeminal neuropathy	39 (78%)	15 (38%)	p<0.01
Facial palsy	0 (0%)	0 (0%)	NS
Hearing loss	48 (96%)	36 (92%)	NS
IX and X cranial nerve signs	7 (14%)	0 (0%)	p=0.02
Cerebellar ataxia	32 (64%)	0 (0%)	p<0.01

\*The proportional difference between the large and small to medium tumours was analysed using the  $\chi^2$  with Yates collection or Fisher's exact probability test.

### Long term follow up and tumour recurrence

The follow up period after the tumour removal was 12 to 115 months (mean 58 (33) months). There were no deaths relating to the acoustic neurinoma during the follow up period. Three patients died from cancer (two of the stomach and one of the pancreas), one from myocardial infarction, and one from an intracerebral haemorrhage. Four patients (8%) had tumour recurrence and underwent a second tumour removal. The interval between the first and second operations was between 27 and 54 months (mean 41 (11) months). The initial tumour removal in these four patients with tumour recurrence was subtotal, and none of 43 patients with total removal had recurrences ( $p<0.0001$ ,  $\chi^2$  test with Yates correction) (fig 5).

## DISCUSSION

### Preoperative characteristics of large acoustic neurinomas

In our series, 22% of patients with large tumours had useful hearing preoperatively, higher than previously reported.<sup>10</sup> Trigeminal neuropathy, the second most common cranial nerve sign, occurs in of 45%<sup>10</sup> to 70% of patients.<sup>20</sup> In our series, the preoperative trigeminal neuropathy usually resolved after tumour removal. Patients with large tumours commonly present with cerebellar ataxia.<sup>20</sup>

Hydrocephalus caused by acoustic neurinoma results not only in symptoms of increased intracranial pressure (headache and papilloedema), but also in symptoms of normal pressure hydrocephalus (gait disturbance, dementia, incontinence) in elderly patients.<sup>21–22</sup> Pirouzman *et al* reported that 36 of 39 patients (92%) with cerebellopontine angle tumour associated with hydrocephalus showed symptoms of normal pressure hydrocephalus.<sup>22</sup> Although tumour removal often causes resolution of hydrocephalus,<sup>23–24</sup> one third of patients required CSF diversion.

**Table 2** Preoperative hearing of 50 patients with large acoustic neurinomas

PTA (dB)	No of patients	Hearing level*	No of patients
<30	4	Class A	4
30–50	8	Class B	7
50–70	11	Class C	12
>70	27	Class D	27

Hearing level\*: classification according to the guidelines of the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology, Head and Neck Surgery Foundation. PTA, pure tone average.

### Retrosigmoid approach in the lateral position

For more than 20 years, we have removed acoustic neurinomas by the retrosigmoid suboccipital approach, with the patient in the lateral position. Many surgeons prefer the sitting/semisitting position for retrosigmoid removal because it is more comfortable for the operator.<sup>6–7, 25–26</sup> However, the semisitting position risks air embolism irrespective of anaesthetic monitoring to prevent this complication.<sup>27–28</sup> In addition, Samii *et al* report a high incidence of haematoma after retrosigmoid removal of cystic tumours in the semisitting position.<sup>6</sup> We removed all the tumours described here with our patients in the lateral position, and no haematomas developed postoperatively. In the semisitting position, the reduced intracranial venous pressure shrinks the peritumoral vein, which prevents troublesome intraoperative bleeding. Thus the vein is likely to be overlooked until a postoperative haematoma develops. In the lateral position, the peritumoral veins cause troublesome intraoperative bleeding, and thus require haemostasis.

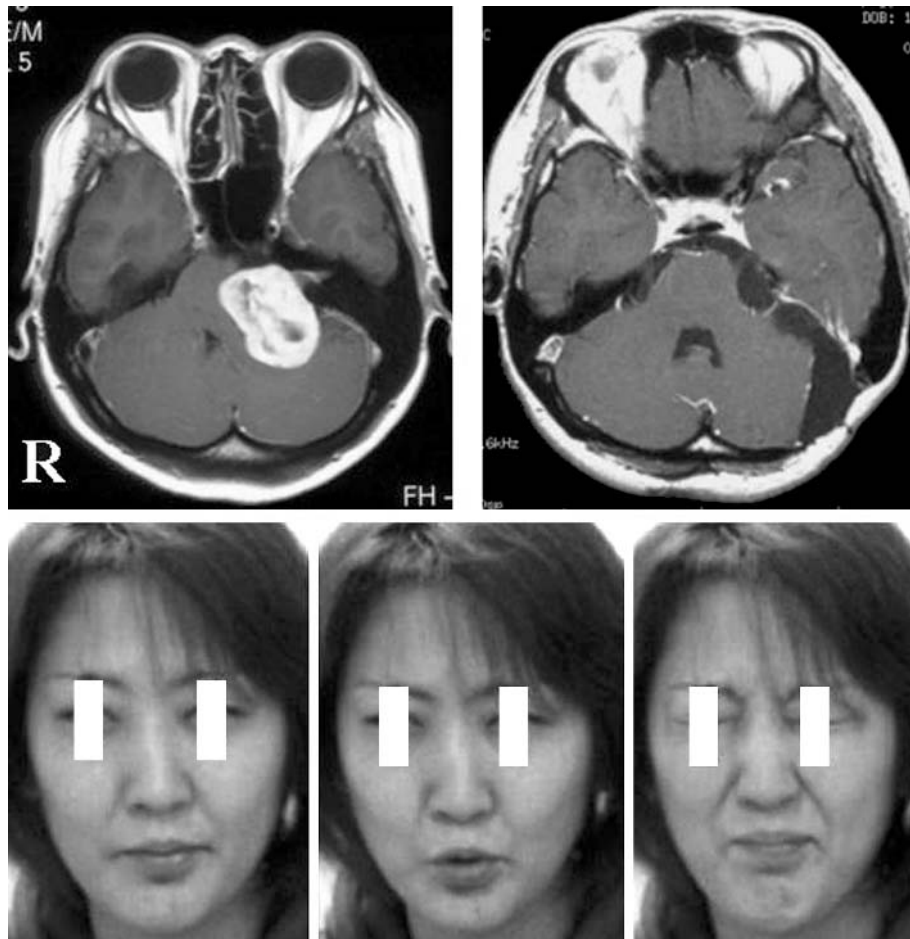
### Completeness of tumour removal

We were able to remove 86% of the tumours completely. As tumour removal was intentionally subtotal—in three patients at the request of the patient—the rate of total tumour removal analysed on an “intention to excise” basis was 92% (43/47). Lanman *et al* report a higher rate (96.3%) of total removal by the translabyrinthine approach.<sup>10</sup> However, the mean tumour diameter in their series was smaller than in ours (3.6 v 4.1 cm), and the percentage of extra large tumours (>4 cm) in their series was also lower than in ours (36% v 50%). Moreover, the incidence of postoperative facial nerve palsy was higher in their series than in ours (47% v 16%). In our series, the subtotal removal rate decreased significantly from the early to the late study period. This is related to the surgeon's learning curve, because all tumours were removed by a single surgeon. The surgeon's operative experience is critical for optimal removal of acoustic neurinomas.<sup>29–30</sup>

**Table 3** Tumour consistency and size of 89 acoustic neurinomas

Tumour consistency	Large tumour (≥3 cm)	Small-medium (<3 cm)
Solid	18	34
Partly cystic	18	4
Mostly cystic	14	1
Total	50	39

Tumour consistency was significantly different between the large and small to medium tumours ( $p<0.0001$ ;  $\chi^2$  test).



**Figure 2** A large acoustic neurinoma in a 27 year old woman was totally removed: preoperative and postoperative magnetic resonance images (upper row). Postoperative face photographs show the facial nerve function of House-Brackmann grade 1 (lower row).

### Facial nerve preservation

The facial nerve was anatomically preserved in 92% of the patients in our series. The anatomical facial nerve preservation rate is 80–90% with the removal of large tumours either by the translabyrinthine approach<sup>4 10–12 31</sup> or by the retrosigmoid approach.<sup>6 20 32</sup> Preservation of facial nerve function has been reported to be 70–80% after the removal of large tumours either by the translabyrinthine approach<sup>4 10–12 31</sup> or by the retrosigmoid approach.<sup>6 20 32</sup> The definition of a functional facial nerve includes both excellent (H-B grade 1/2) and intermediate (H-B grade 3/4) function. The reported preservation rate of excellent function after the removal of large tumours is 42–52.6%.<sup>4 10–12 31</sup> The high rate of preserved facial nerve function in our series (84%) can be attributed to our use of intraoperative monitoring with the facial nerve stimulator-monitor.<sup>13</sup> This monitoring apparatus uses bipolar electrical stimulation in which low amplitude stimulus allows the accurate localisation of the unrecognisably distorted facial nerve. Surgical techniques for preserving facial nerve function include the early identification of the root entry/exit zone and maximum caution in resecting the last lump of tumour in the extrameatal region just outside the porus acusticus.

### Hearing preservation and the surgical approach

The chance of preserving useful hearing is small with the removal of large tumours, and thus many surgeons have recommended the translabyrinthine approach.<sup>4 10–12 31</sup> However, retrosigmoid tumour removal improved the hearing

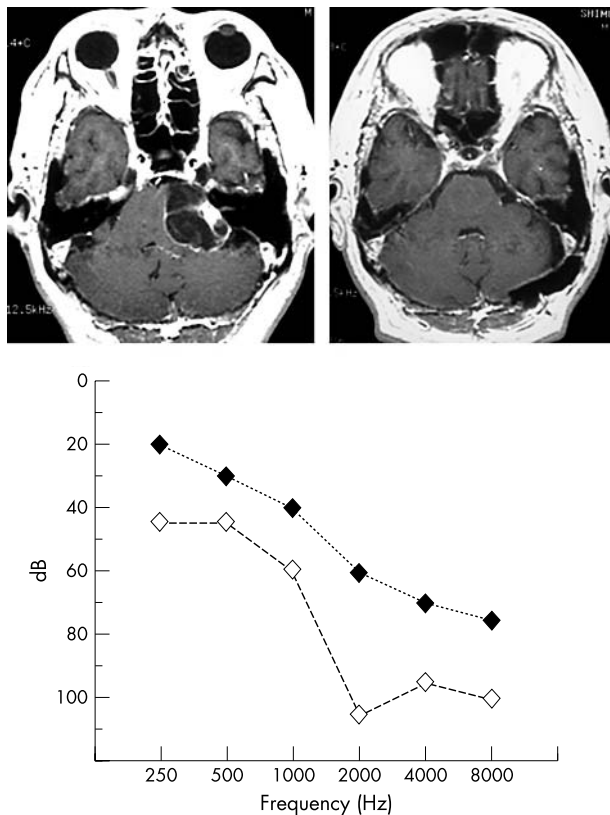
of one patient in our series to a useful level. Using the retrosigmoid approach, Samii *et al* preserved useful hearing in 8% of patients with large tumours who had good hearing preoperatively,<sup>33</sup> and Fahlbusch *et al* retained the hearing of 27.5% of their patients with large tumours who had preserved preoperative auditory brain stem responses (ABR).<sup>34</sup> We routinely monitor the ABR and the cochlear nerve compound action potentials (CNAP) intraoperatively when removing small acoustic neurinomas with the intention of preserving hearing.<sup>35</sup> With most large acoustic neurinomas, ABR potentials are not preserved preoperatively and intraoperative CNAP recording is technically impossible.<sup>35</sup>

### Postoperative complications

Cerebellar and brain stem injuries are the major complications of the retrosigmoid approach, with an incidence of 1.0–2.2%.<sup>6 36</sup> A cerebellar haematoma developed in one patient of our series owing to perforation of the tumour capsule and rupture of the draining vein on the capsule during internal tumour decompression with the ultrasonic surgical aspirator. Ataxia occurs after the translabyrinthine removal of large tumours with an incidence of 12.6%, although the mechanism has not been reported.<sup>10</sup> We observed that preoperative ataxia resolved rapidly after retrosigmoid tumour removal.

CSF leaks and meningitis are the most common complications after the removal of acoustic neurinomas. The translabyrinthine removal of large tumours is associated with a high incidence (14–18%) of CSF leaks.<sup>10–12 37</sup> However, the incidence of CSF leaks was 4% in our series. To prevent

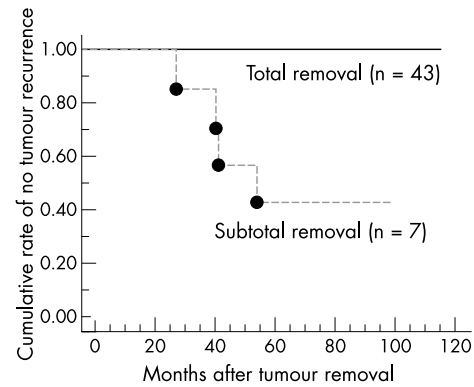




**Figure 3** A 62 year old man who recovered useful hearing after removal of left large acoustic neurinoma: preoperative and postoperative magnetic resonance images (upper panels). The tumour removal improved the pure tone average from 68 dB to 43 dB (lower panel, white symbols, preoperative; black symbols, postoperative).

CSF leaks, we have routinely evaluated the pneumatisation of the temporal bone using preoperative bone window computed tomography (Yamakami I, unpublished data), and have reconstructed the internal auditory meatus.<sup>15</sup> Reconstruction of the meatus prevents CSF leakage through the surrounding air cells. Watertight dural closure and the absence of an epidural or subcutaneous drain prevent CSF leaks. The incidence of meningitis after the translabyrinthine removal of large tumours is 3.7–9.2%<sup>4 10–12 37</sup>; there was no case of meningitis in our series.

Sluyter *et al* reported that aphasia, attributed to left temporal lobe dysfunction, occurred in 12 of 58 patients



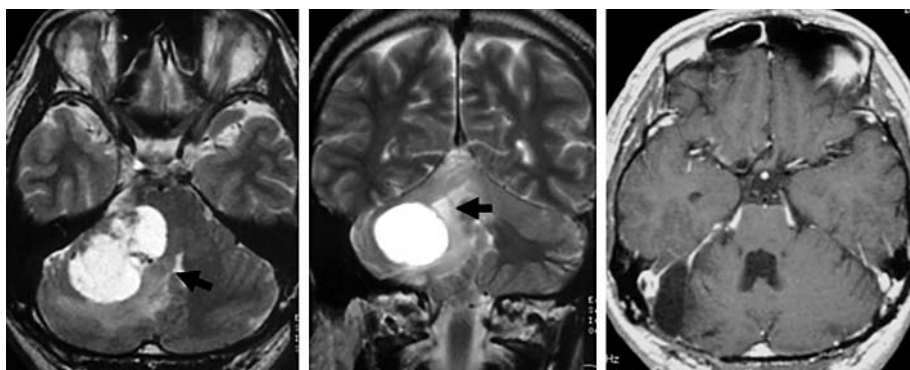
**Figure 5** Cumulative rate of non-recurrence after removal of large acoustic neurinomas; Kaplan-Meier analysis. Tumour recurrence rate is significantly different between total and subtotal removals ( $p < 0.0001$ ).

(21%) after the translabyrinthine removal of large left sided acoustic neurinomas, and that 3.3% of 120 patients had persistent epileptic seizures requiring drug treatment.<sup>11</sup> The venous drainage of the temporal lobe depends on the vein of Labbé and the sigmoid sinus. By compromising these veins,<sup>38–40</sup> translabyrinthine surgery can cause temporal lobe injury. The retrosigmoid approach does not affect the venous drainage from the temporal lobe, and there was no temporal lobe dysfunction in our series.

Postoperative chronic headache has been reported to be a troublesome complication after the retrosigmoid approach.<sup>41</sup> Bone dust generated by the intradural drilling can cause chronic postoperative headaches,<sup>42</sup> so when we drill the posterior wall of the internal auditory meatus, we protect the cerebellopontine angle with pieces of rubber dam and clean out the bone dust by continuous irrigation and suction. We routinely do a cranioplasty to prevent the posterior fossa dura from scarring the suboccipital muscle.<sup>43</sup> No patients in our series complained of chronic headache postoperatively.

#### Long term follow up and tumour recurrence

The tumour recurrence rate after the removal of acoustic neurinomas is 0.4–2%<sup>9 44</sup>; however, the rate after the removal of large tumours has not yet been reported. All patients in our series underwent regular long term follow up with MRI, which showed that the recurrence rate after the removal of these large tumours was 8%. However, all these cases occurred after subtotal removal (four of seven patients), and no recurrence was seen after total tumour removal. The completeness of tumour removal definitely determines tumour recurrence.



**Figure 4** A large acoustic neurinoma with peritumoral oedema of a 52 year old man, which was totally removed: preoperative (left, middle) and postoperative (right) magnetic resonance images. Tumour removal resolved the severe preoperative ataxia as well as the oedema. The peritumoral oedema extended to the middle cerebellar peduncle (arrow).

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Competing interests: none declared

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